CLINICAL PROCEEDINGS

of the

CHILDREN'S HOSPITAL

WASHINGTON, D. C.

September 1954

VOLUME X NUMBER 9

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"FLUSH" TECHNIQUE IN THE DETERMINATION OF BLOOD PRESSURE IN NORMAL INFANTS AND IN INFANTS WITH COARCTATION OF THE AORTA

Ardwin H. Barsanti, M.D.*
Roland W. Penick, M.D.
Bernard J. Walsh, M.D.

Palpation of femoral pulsations in an infant is not only difficult, but the presence of pulsations does not rule out the diagnosis of coarctation. This has been our experience as well as that of many others including Colodny and Carson,[6] who described ten of twenty patients with proved coarctation who had palpable femoral pulsations and stated, "It is evident from these findings that the diagnosis of coarctation of the aorta cannot be excluded solely because femoral pulsations are present."

Coarctation of the aorta is a clinical diagnosis infrequently made in newborns and young infants.[20] Yet, this congenital lesion accounts for a large percentage of infants who develop congestive heart failure and die in the first few months of life.[2] Many of these cases could be saved if the diagnosis were made early enough.

The only accurate method of diagnosis is the demonstration of an abnormal relationship between the blood pressure in the arms and legs, i.e., hypertension in the upper extremities as compared with the lower. It is the purpose of this paper to describe a modification of the "flush" method originally described by Goldring and Wohltmann and to evaluate normal variations in arm-leg blood pressure relationships in young infants. We hope that this will stimulate routine determinations of blood pressure in young infants.

METHOD

All pressures were obtained following as carefully as possible a uniform routine under similar conditions with the child quiet, but not at completely basal conditions, since he usually was being fed. Infants were selected at random, both sexes, negro and white races being included, the only criterion being that they show no evidence of having cardiovascular disease. For the most part, they were under six months, although a few older ones were selected to compare results obtained by the auscultatory method. Two cuffs and two mercury manometers were used. The cuff size was regulated by appropriately folding the cuff when necessary so that it covered approximately two-thirds of the arm or thigh. One cuff was wrapped around the

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hand or foot and the other placed in the usual position around the arm or thigh. The lower cuff was inflated and held at a pressure which would blanch that particular portion of the extremity. The upper cuff was then inflated to a pressure certain to be above that of the systolic pressure. The lower cuff was then removed. While one observer slowly lowered the pressure in the upper cuff, the other carefully observed the blanched portion of the extremity and the systolic pressure was recorded at which a flush was first apparent. The two observers then changed positions and the procedure was repeated so that independent observations were obtained. Two readings were obtained in the upper extremity and in the lower extremity in each case. If the child was irritable, various time-honored measures were employed in an attempt to quiet him. Feeding was found to be most effective. This allowed a stable level of blood pressure to be obtained which usually did not vary more than 2–6 mm. Hg between the two observers. The use of the second cuff to blanch the extremity (hand or foot) was found most effective because of its relative ease and rapidity and with a minimal chance of disturbing the infant.

RESULTS

As is seen from charts I–IV (range) the values obtained by two observers are fairly close, usually between 2–6 mm. Hg (larger discrepancies usually indicate an irritable infant). This points out the relative accuracy and reproducibility of this fairly crude procedure. This probably is due to the

CHART I

Under One Week

No.	Age	Arm		Leg		Arm-leg Difference
		Average	Range	Average	Range	
Group I: Arm pressure exceeds leg pressure*						
1	2 days	70	68–72	72	70–74	2
2	2 days	81	80–82	80	80–80	1
Average difference						1.5
Group II: Leg pressure exceeds arm pressure						
1	2 days	70	68–72	72	70–74	2
2	1 day	61	60–62	67	66–68	6
3	1 day	70	70–70	74	72–76	4
4	5 days	83	80–86	86	84–88	3
5	5 days	82	80–84	83	82–84	1
6	3 days	73	72–74	75	72–78	2
Average difference						3

* All pressure readings are given in millimeters of mercury.

CHART II

Under Three Months

No.	Age	Arm		Leg		Arm-leg Difference
		Average	Range	Average	Range	
Group I: Arm pressure exceeds leg pressure						
1	2 weeks	70	68–72	59	50–68	11
2	2 weeks	87	86–88	79	78–80	8
3	3 weeks	91	88–94	86	84–88	5
4	3 weeks	89	88–90	88	88	1
5	1 month	83	84–82	81	82–80	2
6	2 months	99	98–100	95	94–96	4
7	2 months	90	90	87	86–88	3
8	2.5 months	111	110–112	109	108–110	2
Average difference						4.5
Group II: Leg pressure exceeds arm pressure						
1	2 weeks	59	58–60	69	68–70	10
2	1 month	106	102–110	110	100–120	4
3	1.5 months	105	104–106	106	106	1
4	1.5 months	90	88–92	92	92	2
5	2 months	88	82–94	100	96–104	12
6	2 months	101	98–104	104	102–106	3
7	2 months	81	80–82	107	106–108	26
8	2.5 months	64	62–66	66	64–68	2
9	2.5 months	105	104–106	109	104–114	4
10	2.5 months	106	104–108	113	112–114	7
Average difference						6.6

rather sharp end point or "flush" seen in these small infants as compared to the more gradual spread of the flush in older children. It therefore constitutes a more accurate procedure in the age group where the blood pressure is most difficult to obtain by the conventional auscultatory method.

The area of confusion lies in those cases where the pressure in the upper extremities is found to be higher than in the lower extremities. As can be seen from our results, the pressure in the arm can normally be as much as 12 mm. Hg higher than in the legs. Our impression was that this was usually found in the younger infants or the pressures tended to be equal in these infants. However, an insufficient number of determinations in older infants were made to conclusively demonstrate this impression. In no cases, was a pressure gradient of more than 15 mm. Hg noted in the arm over the leg.

In the determination of the blood pressure level per se, it is necessary

CHART III

Three to Six Months

No.	Age	Arm		Leg		Arm-leg Difference
		Average	Range	Average	Range	
Group I: Arm pressure exceeds leg pressure						
1	3 months	111	110–112	100	98–102	11
2	3 months	96	96	94	92–96	2
3	4 months	76	70–82	69	68–70	7
Average difference						10
Group II: Leg pressure exceeds arm pressure						
1	3 months	119	118–120	131	130–132	12
2	3 months	103	102–104	107	106–108	4
3	3 months	95	94–96	107	106–108	12
4	3 months	102	98–106	104	98–110	2
5	3 months	78	78	86	86	8
6	3.5 months	117	116–118	130	128–132	23
7	3.5 months	113	114–112	125	124–126	12
8	4 months	94	92–96	97	96–98	3
9	4 months	99	96–102	103	102–104	4
10	4 months	100	98–102	106	106	3
Average difference						8.3

for the infants to be as close to basal conditions as possible, since the slightest activity, such as sucking, will cause elevation of the blood pressure and marked variations between consecutive determinations. However, this is not particularly important, since it is the relationship between the arm and leg in which one is interested in attempting to rule our coarctation.

The following two cases are presented to illustrate the importance of this procedure:

Case # 1

J. B., white female newborn, was well until four days of age when tachypnea and fever developed. Fever subsided but dyspnea and retraction persisted and periods of cyanosis were noted by the parents. On admission at ten days of age, dyspnea and retraction were noted. There was a precordial bulge, the liver extended down to the umbilicus. No murmurs were heard. The lungs were clear. Femoral pulsations were felt; however, blood pressure in the upper arm was 120 mm. Hg and in the leg it was 40 mm. Fluoroscopy revealed enlargement of both ventricles. Electrocardiogram revealed definite right ventricular hypertrophy. The infant's condition became progressively worse, in spite of oxygen and digitalis, and she died at eighteenth day of age.

At autopsy, a preductal coarctation was found with patent ductus arteriosus and marked right ventricular hypertrophy.

CHART IV

Six to Nine Months

No.	Age	Arm		Leg		Arm-leg Difference
		Average	Range	Average	Range	
Group I: Arm pressure exceeds leg pressure						
1	6 months	121	120–123	109	108–110	12
2	7 months	106	106	105	104–106	1
3	8 months	110	104–116	107	104–110	3
Average difference						5.3
Group II: Leg pressure exceeds arm pressure						
1	6 months	87	86–88	89	84–94	2
2	6 months	72	70–74	73	72–74	1
3	6 months	78	76–80	80	78–82	2
4	6 months	93	92–94	97	96–98	4
Average difference						2.2

CHART V

Nine to Fifteen Months

No.	Age	Arm		Leg		Arm-leg Difference
		Average	Range	Average	Range	
Group I: Arm pressure exceeds leg pressure						
1	10 months	100	98–102	91	88–94	9
2	15 months	93	88–98	87	84–90	6
Average difference						7.5
Group II: Equal pressure in arm and leg						
1	15 months	99	98–100	99	98–100	0

Case # 2

D. B., an eight and a half year old white male, was admitted for operation with a diagnosis of coarctation of the aorta. Blood pressure in the upper extremities (auscultation) was 190/90 and was unobtainable in the lower extremities. By "flush" technique the blood pressure in the arm was 180, and in the legs 80 mm. Femoral pulses were not palpable.

At operation an area of coarctation about 2.5 cm. long was found just distal to the entrance of the ductus arteriosus. The dorsalis pedis and femoral pulses were palpable for the first time 72 hours post-operatively. Blood pressure at this time was 150/98 in the arms and 130 in the legs by auscultation, and 110 using the "flush" technique.

In this case, the "flush" technique allowed for a more accurate post-operative follow-up.

DISCUSSION

There have been numerous studies dealing with the normal variations of blood pressure in children and adults.[18, 16, 11, 8, 10, 19] Most of them deal with older children where the auscultatory method is used.

The normal variations with respect to age have been the subject of several papers.[9, 12, 14, 17, 22, 26] The selection of the proper size cuff has been considered most important. Woodbury, Robinson and Hamilton measured the intra-arterial pressure in newborns and found that the 2.5 cm. cuff would be most accurate in this age group. Robinson, Hamilton, Woodbury and Volpitto[22] in a study of 62 children from six weeks to thirteen years felt that the width of the cuff should increase with age. They concluded that a 2 cm. cuff for newborns, a 5 cm. cuff for infants under one year, and a 9 cm. cuff for those over one year correlated most closely with intra-arterial pressures taken at these ages. Day,[10] on the other hand, in a study of 41 patients, taking simultaneous pressures with an adult cuff on one arm and varied sizes on the other, concluded that the most accurate method consisted of covering the entire upper extremity without infringing on the elbow or axilla. Baranton[3] similarly concluded that the 4 cm. cuff was practical up to six months, but thereafter the width of the cuff should cover the entire upper arm. Downing,[11] on the basis of many observations on girls between the age of 3–16 years, attempted to set forth the ages at which different sized cuffs should be used. Graham et al.[14] felt that a good method was to choose a cuff whose width covered about two-thirds of the extremity. This has been adopted by most and was used throughout our study. If the exact cuff is not available, a larger cuff can be appropriately folded and will give comparable results.[9, 25] We felt that this provided a very convenient and uniform method of study.

It was apparent from a close scrutiny of the literature that no study has been made regarding the normal relationship between the blood pressures in the arms and legs in infants. Studies in older age groups have been carried out. Hamilton, Woodbury and Harper[15] found by direct measurement of intraarterial pressure, that the pressure increases as the distance from the heart increases. Kotte et al.[18] similarly found that the "direct" femoral blood pressure was significantly higher than the brachial in half of their cases. In the others, the difference was slight. Using a photoelectric device, Burdick et al.[5] found that the leg-arm difference in normal males was 20–40 mm. Hamilton and Dow,[16] in an effort to explain this phenomenon, stated that the pressure exerted in a peripheral vessel was a function of the kinetic energy from each systolic contraction. The kinetic energy in turn is a function of the mass and velocity of the blood as expressed:

$$(\text{K.E.} = \tfrac{1}{2}(\text{Mass} \times V^2))$$

The longer the blood vessel, the greater the blood mass, the greater will be the K.E., and therefore the lateral pressure. In addition to this, Wendhos et al.[24] in a study of blood pressure in the lower extremities in 500 normal males, found a direct correlation between the circumference of the extremity and the height of the blood pressure. They postulate that the soft tissue mass is an additional factor in producing a higher blood pressure.

This normal finding of a higher blood pressure in the legs is not so apparent in infants as is demonstrated by our series as well as that of Goldring and Wohltmann.[13] It is therefore of the utmost importance to establish the normal variations in the leg-arm relationship in infants in order to better evaluate abnormal conditions such as coarctation of the aorta. Brown et al.[4] in a study of adults with proved coarctation, found that the average gradient between the arm and leg was 83 mm. In no cases was it less than 20 mm., nor were any normals found where the arm to leg gradient was over 15 mm. This was confirmed in our series of cases as well as in the cases of proved coarctation, J. B. and D. B. (cases 1 and 11). It would be reasonable then to arbitrarily set the normal upper limits for the arm to leg gradient at 20 mm., as has been suggested by Goldring et al.

CONCLUSIONS

In a study of fifty infants, blood pressure readings were taken in the arms and legs using the "flush" technique. The blood pressures were taken in duplicate by two observers.

We have attempted to show, in addition to the description of the method, that:

1. The "flush" technique is practical and accurate in those infants where an auscultatory blood pressure is difficult to obtain.

2. Certain normal variations exist in the arm-leg gradient in infants. Where the arm-leg difference exceeds 15–20 mm. coarctation of the aorta should be suspected.

3. The mechanism for the normal leg-arm gradient is briefly discussed.

4. Proper medical measures and advancing surgical technique can save infants who otherwise would die of coarctation in the first few months of life. Increased awareness of this, coupled with more frequent routine determination of blood pressure in both arms and legs in these infants, can provide a more rapid diagnosis and early treatment.

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CHONDRODYSTROPHIA CALCIFICANS CONGENITA (STIPPLED EPIPHYSES)

Belinda Straight, M.D.

Chondrodystrophia calcificans congenita is a rare disease found in infants and small children, which is diagnosed by the stippled appearance of the epiphyses on x-ray, contractures of the joints, shortening of one or more limbs and congenital cataracts. Conradi[5] first described this condition in 1941. He reported a case with numerous calciferous opacities within the cartilage of the epiphyses. These opacities disappeared without treatment, and he considered it a form of chondrodystrophy. Since this time some thirty-seven cases have been reported.

CASE REPORT

This five day old colored male baby was born at Garfield Memorial Hospital on October 15, 1953, and was diagnosed as having chondrodystrophia calcificans con-

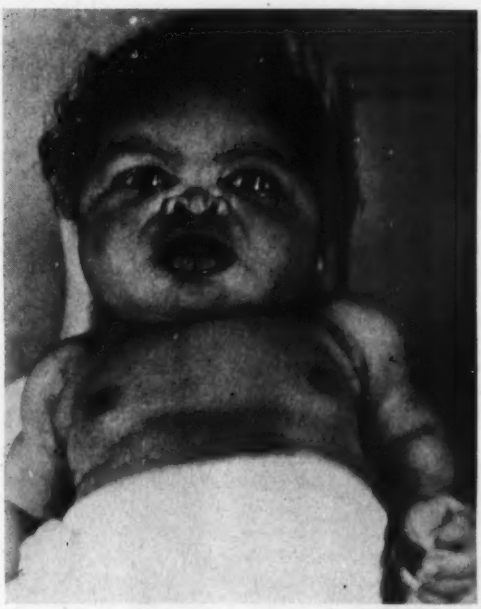

FIG. 1. Photograph of patient showing characteristic facies, short neck, barrel-shaped chest, and flexed upper extremities.

genita by x-ray on the second day. On the fourth day he developed an abscess of the scalp and was admitted to Children's Hospital.

The mother was a primagravida, Rh negative, group B, with no rise in antibodies during pregnancy. She had malaria during the first trimester of pregnancy. Labor was 21 hours in duration with a persistent posterior presentation, rotated with forceps. At birth it was noted that the baby had a wide face with fat cheeks, absent external nose, with flaring nostrils. The infant was dyspneic, but nursed well from the breast and cried lustily with deep harsh note. On the fourth day he developed a fever and on the fifth there was a soft area, measuring 2 cm in diameter, over the right parietal region from which thick yellow pus could be expressed.

The family history was noncontributory other than malaria in the mother during the third and fourth months of pregnancy. This was treated with quinine. There were no other members of the family who had deformities of the bones or face. Unfortunately no x-ray examinations of the parents were obtained.

Physical examination on admission to Children's Hospital revealed a large, active baby, weighing 4.41 kg. with absent bridge of nose and its cartilage, fat cheeks, and hirsutism of the forehead. The head measured 37.5 cm. in circumference. The anterior fontanel admitted two fingers, the posterior, one. There was a hard prominence over the left parietal area which measured 3 cm., in diameter. Fundoscopic examination of the eyes revealed a discoid thickening of the lens; the optic nerve head was pale, narrow and incompletely oval. The vitreous and corneae were clear. The blood vessels at the discs were larger than normal. Most of the retinae was seen inferiorly. They

were grey in color. This picture was consistent with early retrolental fibroplasia, but no hemorrhages or exudates were seen. Optic atrophy was the primary diagnosis.

The nose was described. Both passages were open. There was a nasal septum. The mouth had a normal palate with a short uvula. The neck was short. The head appeared to arise from the chest, which was barrel-shaped. The lungs were clear. The heart had regular sinus rhythm and no murmurs were heard. The cord was dry, the abdomen soft, the liver being palpated two fingerbreadths below the right costal margin. No other masses or organs were felt. The testes had not descended.

The legs were hypotonic and could be easily extended above the head. The upper extremities had limitation of motion, particularly on the right. The right elbow was flexed, the patient crying when it was extended. The toes were short and stubby. The skin was clear except for the abscess of the scalp. On neurological examination the Moro reflex was sluggish.

The baby's blood was Rh positive, group A. There was a negative Coombs test on the second day. The serology tests of mother and baby were negative. On October 15, 1953 the hemogram was as follows: Hemoglobin 13 gm., 4.5 million erythrocytes and with 28 nucleated red cells/100 leukocytes. On October 17, 1953 the hemoglobin was 17 gm. with 4.5 million erythrocytes and 1 nucleated red cell/100 leukocytes. Urine on

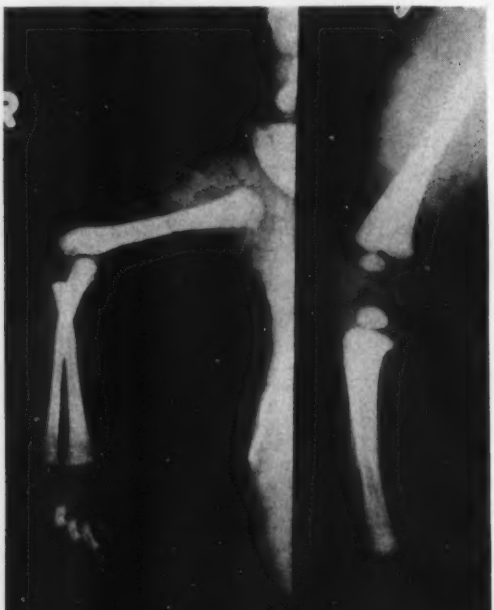

FIG. 2

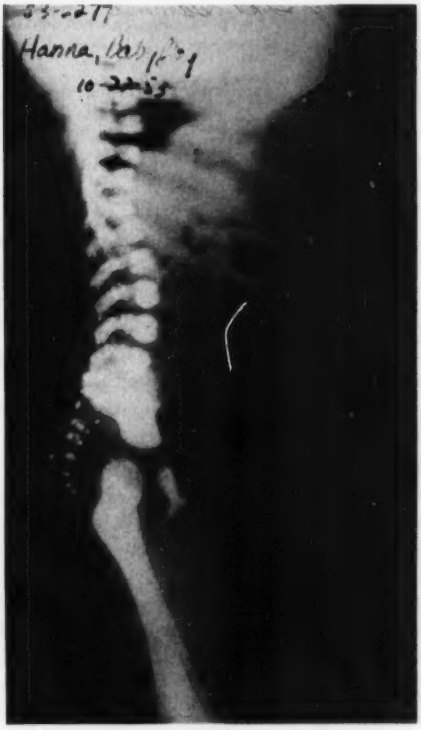

FIG. 3

admission to Children's Hospital had 20–30 leukocytes/hpf, with many finely granular and coarsely granular casts and 50 mg./100 ml. of albumin. The urine returned to normal within a week.

Blood chemistries which included sodium, potassium, calcium, phosphorus, alkaline phosphatase, total protein, albumin, globulin, a/g ratio, urea nitrogen and serum cholesterol, were all within normal limits for the patient's age. Cultures taken from the infected area of the scalp grew out hemolytic *Micrococcus pyogenes, var. albus*.

Roentgenograms were reported as follows: "The skull shows a wider than usual calvarium. The fontanelles are large but not bulging. A metopic suture is present. There is no external nose visible. There is stippling of the manubrium, the epiphyses of the head of each humerus, the head of each femur, of the sacrum, coccyx, the tarsal bones and the left acetabulum. There are stippled epiphyses along the lower dorsal and lumbar spines. There is a left dorsal lumbar scoliosis. The chest is not remarkable. There are speckled areas of mineralization in the left elbow. Both elbows are congenitally dislocated. There are lime deposits in the knee joints and in the right hip. There is deposition of calcium salts in the soft tissues surrounding the right shoulder joint."

The patient received 150,000 units of penicillin, twice a day intramuscularly and saline soaks to the scalp. The furuncle of the scalp improved and he was discharged on the seventeenth hospital day.

At two months of age the patient was eating well, he had gained weight, and showed no obvious retardation. He developed an atopic dermatitis and was put on a soy bean formula and is now being followed in the skin clinic. He was seen at the plastic surgery clinic, where plans were made for future surgery.

DISCUSSION

Chondrodystrophia calcificans congenita was given its name by Raap [11] in 1943. Other names include hypoplastic fetal chondrodystrophy, dysplasia epiphysialis punctate, stippled epiphyses, and calcinosis universalis.

The etiology is unknown. The frequency of parental consanguinity is higher (probably over 12 per cent) than that of the general population (about 10 per cent). Frazer and Scriver point out that of 33 reported families six contain more than one case. They demonstrate a simple mendelian recessive inheritance in this disease.

The role of nutrition in the mother has not been fully investigated, nor have sufficient blood chemistry studies been done on these mothers immediately after delivery. Although this patient was the product of a pregnancy accompanied by febrile illness, such is not the case with most of the patients who have been reported.

Briggs et al.[2] give the pathologic findings as follows: "The most striking feature is the variation in cellular concentration, the great increase in vascularity of the cartilage, and areas of apparent degeneration and primary calcification. The vessels appeared to penetrate into the cartilaginous mass from the immediate joint surface." Fairbank[6] speaks of the pathology as being one of mucoid degeneration of the epiphyseal cartilage, with calcification following this. Calcification of the synovial tissue is described by Borovsky and Arent.[3]

The chief complaint may be the abnormal facial appearance, stiffness of the joints and inability to straighten out the limb. These evident abnormalities have led to early x-ray diagnosis. Several children mentioned in the literature have deformities of the nose. More common are bilateral congenital cataracts and or optic atrophy. A short neck may occur. Mental retardation is one of the hazards of the disease. Congenital heart disease has been described, and so has hydronephrosis with hydroureter. Limitation of motion of the extremities may be accompanied by dislocation of the hips or the elbows.

There is a frequent complaint of poor weight gain and feeding difficulties. Resistance is poor. About half of the babies die during the first year of their lives. Death usually comes from urinary tract infections, tuberculosis, bronchopneumonia, or cardiac failure.

Diagnosis is made by x-ray examination. Stippled discrete calcifications appear in the epiphyseal regions. These are found wherever cartilage is normally found. They commonly appear at the proximal and distal ends of the femur and the proximal ends of the tibia and the humerus. The tarsal and carpal areas may be involved. The hyoid bone, transverse processes, epiphyses for the end of the ribs, and sacrum are sites for such lime deposits. Fairbank's[6] description of the x-ray appearance remains a classic, viz., "Suggestive of that produced by flicking paint from a brush to a clean surface."

Premature calcification occurs in areas never ossified at birth, and this does help to distinguish the stippling from that of hypothroidism. Ossification in the latter is delayed. Vinke[12] feels that chondrodystophia calcificans congenita should be differentiated from stippling in older children which does not occur within the cartilage.

Shortening of the shafts of long bones may be seen on x-ray. The metaphyses tend to flare. Dwarfing is of the short limb kind, the proximal limbs being more involved than the distal in the disease of the three C's.

In most of the cases reported the laboratory studies were normal, although elevated blood calcium and serum phosphatase have been recorded.

The prognosis is guarded. One half of the children die within the first year. Others may have some mental retardation, blindness, or dwarfism. Caffey,[4] however, feels that the prognosis for complete recovery without residual deformities is good provided that the infantile period is survived. In these patients the lime deposits decrease and normal bone development appears to replace them.

SUMMARY

A case of chondrodystrophia calficans congenita in a negro baby is presented. The patient shows many of the classical features of the disease. The absent external nose, optic atrophy, short neck, congenital dislocation of the elbows, flexion contractures and roentgen appearance all serve to confirm the diagnosis. The normal blood chemistries are consistent with these findings in previously reported cases. No familial trend in this patient was noted. The history of malaria in the mother in the first trimester of pregnancy might possibly have been of some significance.

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GROSS HEMATURIA IN A CHILD WITH SICKLE CELL ANEMIA

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A. T., a nine year old colored boy, was admitted to Children's Hospital on October 31, 1953 because of massive, painless hematuria. The child was known to have sickle cell disease. He was apparently well until the day prior to admission, during the morning of which he complained of mild upper abdominal pain, which lasted a few hours and which disappeared spontaneously. Later in the day the mother noticed bloody urine and on one occasion blood clots were seen in the urine. The child stated that no hematuria had occurred previously. There was no discomfort associated with urination and there was no nocturia or renal pain. No bleeding from any other site was observed. Appetite and general condition were thought to be excellent. He had been afebrile.

PAST HISTORY

This was the patient's forty-seventh admission to this hospital. He was first admitted at the age of nine months because of an upper respiratory infection and at that time it was found that he had sickle cell disease. Subsequent admissions were usually for the treatment of respiratory infections or sickle cell crises. In the course of nine years the patient received a total of 24,000 ml. of blood at this hospital (Table 1).

A splenectomy was performed in February, 1949 with no appreciable effect on the course of his disease. His growth and development have otherwise been normal.

Daily urine analyses during this admission in 1949, were normal. The specific gravity varied from 1.000 to 1.024. No albumin was found and only an occasional leucocyte was seen in the sediment. No blood clots were observed in the urine specimens.

Family History: Father, mother and five siblings are living and well. There was no evidence of sickle cell anemia in the other members of the family.

Physical examination on admission revealed a happy and active colored boy in no

TABLE 1

Year	Amount of Whole Blood	Number of Transfusions
	ml.	
1945	1275	9
1946	2450	13
1947	1650	8
1948	3250	12
1949	3500	13
1950	4050	13
1951	3250	9
1952	2925	11
1953	1650	5
Total.....	24,000	92

TABLE 2

Day of Hospitalization	Color	Sp. Gr.	Alb. mg/100 ml.	RBC	WBC	Sediment-centrifuged per. H.P.F. Casts—Crystals
1	Bloody	1.012	80			The sediments contained erythrocytes, many of which were sickled. No casts seen
2	Bloody	1.013	200			
3	Bloody	1.012	200			
4	Bloody	1.015	100			
5	Bloody	1.014	200			
6	Bloody	1.017	200			
7	Bloody	1.016	200			
8	Cloudy	1.016	Trace	5–10		
9	Hazy	1.016	—		1–2	
10	Clear	1.014	—	—	—	

acute distress. The temperature was 100.2° F (rectal), pulse 88 per minute, respirations 24 per minute and blood pressure 110/65.

The sclerae were icteric and the mucous menbranes were pale. There was a mild generalized lymphadenopathy. The heart was slightly enlarged on percussion and a grade 2 systolic murmur could be heard over the apical area. The liver edge was palpable 5–6 cm. below the right costal margin and it was sharp, smooth, firm and non-tender. There was no costovertebral or flank tenderness and the bladder area could be palpated without causing pain. The external genitalia were normal. The examination was otherwise essentially normal. Table 2 shows results of the work-up done during this admission:

Urine Cultures: No growth on several occasions.

Cystoscopy performed on November 17, was essentially negative except for minimal trabeculation of the bladder.

X-ray Studies: (Dr. I. Lattman) Flat film of the abdomen. "The kidneys are well visualized as are the psoas muscles. There are no shadows suggesting urinary calculi. Regrograde studies of both urinary tracts show no gross abnormality of the calyces, pelves or ureters. Fine calyceal detail is lacking. The kidneys are normal in outline."

Day of Hospitalization	HGB Gm/100 ml.	WBC	Seg.	Band	Young	Lymph.	Mono.	Eosin.	Baso.	Platelets
1	7.5	17.3	28			46	1	25		Normal
4	8.5	18.9	51			25		24		Normal
5	7.8	22.1	68			15		17		Normal
6	8.5	18.7	42	3		27		27	1	Normal
11	8.5	13.3	58	5		36		1		Normal
19	9.0	14.4	66	3	1	22	1	7		Normal
20	5.8	14.4	64	15		17	3		1	Normal

Direct Coombs Test: Negative.

Bleeding time, coagulation time and prothrombin time were normal.

Serological tests for syphilis: Negative.

Several sickle cell preparations (sodium metabisulfite method) done on different days, showed 50–90% sickling.

Blood urea nitrogen: 8 mg/100 ml.

No cold agglutinins were demonstrated.

Bone marrow examination showed erythroid hyperplasia, marked granulophilia and normal platelets. No megakaryocytes were seen.

Stools: Repeated examinations for ova and parasites were negative.

Liver Function Tests: No bilirubinuria. Thymol turbidity: 2 units. Cephalin flocculation: Negative. On day of admission: Direct bilirubin: 0.6/100 ml. Indirect bilirubin: 1.10 mg./100 ml. On fifteenth day of admission: Direct bilirubin: 0.10 mg./100 ml. Indirect bilirubin: 0.86 mg./160 ml.

Course in Hospital: The abrupt cessation of the hematuria, which lasted a total of eight days, was striking. The child had several episodes of severe abdominal pain during his stay in the hospital, both before and after the cessation of the hematuria. He received a blood transfusion on the twentieth hospital day at which time a drop in hemoglobin, accompanying an attack of abdominal pain occurred. This was indicative of a sickle cell crisis.

The patient was discharged on November 29, 1953 to be readmitted in January 1954, at which time the history and physical findings were similar to those reported at his previous admission. This time the massive hematuria lasted six days and again stopped abruptly. The following are the results of the workup done during this admission:

Urological Examination:

Urinalyses

Day of Hospitalization	Color	Sp. Gr.	Alb. Mg%	RBC	WBC	Sediment-centrifuged-per H.P.F. Casts—Crystals
1	Bloody	1.028	500			The sediments of these urines contained numerous red blood cells, many of which were in the sickling state.
2	Bloody	1.019	250			
5	Bloody	1.016	500			
7	Clear	1.019	Negative	1–2	Rare	

Repeated urine analyses during the remainder of the admission were normal in every respect.

Excretory Pyelogram: (Dr. I. Lattman) "Examination of the genito-urinary tract after the injection of dye, at the end of 4, 10, 20 and 30 minutes reveals the kidneys, ureters and bladder to fill satisfactorily and to appear normal."

Cystoscopy (Dr. P. Bender) on January 29, 1954: "A number 14 McCarthy cystoscope passed without difficulty. Urine is clear, although it was reported to be bloody yesterday. Bladder is perfectly normal except for pallor of mucosa. Ureteral orifices difficult to visualize. Unable to pass catheters because of buckling."

Blood Examination

Day of Hospitalization	HGB Gm/100 ml.	WBC	Seg.	Band	Lymph	Mono.	Baso.	Platelets
1	6.5	24.6	36	4	52	1	1	Normal
5	6.1	12.2	76		11	5	1	Normal

Bleeding time, coagulation time, prothrombin time, cold agglutinins: normal.

Circulating eosinophils: 310 per mm^3.

Liver function studies: Essentially as on admission in November, 1953.

Blood urea nitrogen: 9 mg./100 ml.

Course in hospital was relatively uneventful.

Following this admission the patient has had two brief admissions because of sickle cell crises, which were manifested by abdominal and joint pains. At neither of these admissions were red cells found in the urine sediment.

DISCUSSION

Sickle cell disease, both in the homozygous form (sickle cell anemia) and the heterozygous form (sickle cell trait or sicklemia) are recognized as a possible cause per se of unexplained gross, renal hematuria. So far about 40 cases have been reported. All of these episodes occurred in adults.

Abel and Brown reported an adult negro patient, who underwent a nephrectomy because of a kidney-tumor was suspected. No sickling could be elicited in the peripheral blood. However, in the microscopic sections of the removed kidney, sickled red blood cells were seen.

Goodwin et al., were the first to suggest sickle cell disease in itself to be a cause of gross hematuria. Microscopy of sections of a kidney removed from a nine year old negro showed, "In some areas the surface of the papillae were partially denuded of epithelium. The underlying stroma showed moderate infiltration and fibrosis. A striking feature of the papillae was the presence of numerous greatly dilated capillaries and venules just beneath the epithelium. There were some vessels filled with red blood cells in the sickling state." The third case in Goodwin's series was a 11-year old boy.

Most reports do not mention any pain of a colicky nature in their clinical histories, but the bleeding being of renal origin, a picture of ureteral colic can develop.

Creedy et al. stress that gross hematuria may be the only manifestation of sickle cell disease.

Harrison and Harrison report nine patients of whom nephrectomy was performed on six. Most authors agree that the only indication for nephrectomy should be bleeding which is not controllable by repeated blood transfusions.

Lund et al. describe seven adult patients with the sickle cell trait who showed gross hematuria. Nephrectomy was performed in two. The percentage of sickling in the peripheral blood varied from 0–80 per cent. With one exception, all bleeding was unilateral, usually coming from the left side.

SUMMARY

Two episodes of massive, painless hematuria occurred in a nine year old boy with sickle cell anemia. Complete urological examination was negative on both occasions. A review of the literature is presented. Gross hematuria may be the only manifestation of sickle cell disease in children.

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ANNULAR PANCREAS IN A PREMATURE WITH OTHER CONGENITAL ANOMALIES

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INTRODUCTION

Annular pancreas is a very rarely encountered anatomical abnormality. In all, only about 85 cases have been reported in the world literature. The majority of these cases have been found at autopsy, and it has been only in recent years that this diagnosis has been either considered clinically, or established operatively. Only a minority of these cases have occurred in children.

Anatomically, as the name implies, annular pancreas consists of a ring of pancreatic tissue which encircles the second portion of the duodenum and which is inseparable from the main portion of the gland, which is otherwise normal. The ductal system in the annulus is variable, but the usual arrangement consists of drainage of the ring by a duct which begins anteriorly and passes to the right and then posteriorly to drain into the main pancreatic duct of Wirsung. Although uncommon, a duodenal stenosis or atresia may be present as an associated abnormality.

The pancreas arises embryologically as dorsal and ventral buds from the endodermal lining of the gut in embryos of 3 to 4 mm. The dorsal pancreatic anlage develops just cephalad of the level of the hepatic diverticulum and grows rapidly into the dorsal mesentery. Unequal growth of the duodenal wall, and elongation of the primitive bile duct cause a rotation of the ventral pancreas laterally to the right and then posteriorly where it fuses indistinguishably with the dorsal anlage.

Majority opinion favors the theory that an annular pancreas is produced when the tip of the ventral pancreatic head becomes adherent to the duodenal wall anteriorly and then, when the normal rotation of the base of the ventral pancreas occurs, a band of pancreatic tissue is dragged around the circumference of the duodenum, thus encircling this structure.

Other theories include that of hyperplasia of the normally situated pancreas as a regenerative response to a fetal peritonitis. The hyperplasia is thought to dissect under the serosa of the duodenum and thereby surround the second portion of this organ. A somewhat similar theory attributes the anomaly to an overgrowth of ducts of the pancreas which are directed circularly, due to the local mechanics.

Pathologic changes found in annular pancreas include pancreatitis, obstruction of the duodenum with dilatation and ulceration of its mucosa, and inflammatory processes of the biliary system. In addition to these local pathologic changes, associated anomalies are found in 25 per cent of cases, according to Cattell. Gross reports an even higher incidence, for nine out of ten of his operated cases had another congenital defect including congenital heart disease, tracheoseophageal fistula, imperforate anus, mongolism, and malrotation of the bowel.

The clinical picture produced by annular pancreas will vary according to the degree of obstruction of the bowel, biliary tract or pancreatic duct. No symptoms referable to this condition may manifest themselves during life and the annular pancreas will be detected as an incidental autopsy finding. However, when obstruction of the duodenum is present, the clinical picture of a high bowel obstruction becomes apparent. In pediatric practice this condition often presents as persistent vomiting in the neonatal period. The vomitus may or may not contain bile, depending upon the level of the obstruction. Classically, vomiting starts following the first feeding and will persist unless the obstructing lesion is removed. An annulus which does not completely compress the lumen of the duodenum will produce a picture of intermittent upper bowel obstruction.

The diagnosis of annular pancreas in the infant will rest on the clinical picture of persistent vomiting and upon x-ray findings. In the classical case, a flat film of the abdomen will reveal a massively dilated stomach and first portion of the duodenum. A lateral film of the abdomen will aid

in the recognition of the dilated duodenal bulb. Little or no gas or fluid will be seen in the remainder of the bowel. These findings are diagnostic of obstruction of the second portion of the duodenum, and can be caused by annular pancreas, duodenal stenosis or atresia, and malrotation of the colon with the nondescended cecum compressing the duodenum.

In case of the incomplete block, flat plate of the abdomen may not be diagnostically significant. An upper gastro-intestinal series will then reveal the characteristic symmetrical smooth narrowing of the second portion of the duodenum.

In the past, surgical attack on an annular pancreas was accomplished by division of the pancreas, but this procedure proved disappointing due to the development of pancreatic fistulas, the possibility of overlooking an associated duodenal stenosis and the possibility of subsequent fibrosis with recurrence of duodenal obstruction.

Gastroenterostomy was next performed; this procedure relieved the obstruction but sometimes failed to allow drainage of the proximal duodenal pouch.

Present opinion strongly favors the employment of a duodenojejunostomy which will effectively relieve the obstruction and which will avoid the cited operative hazards.

We are presenting this case as an interesting example of this unusual anomaly which caused symptoms in a premature newborn and was associated with other congenital defects.

CASE REPORT

The patient was a 5 day old white girl, who was admitted from the hospital of her birth on July 19, 1954 because of a large amount of mucus in the nose and throat, intermittent bile stained vomitus, and roentgen evidence of dilatation of the stomach and first part of the duodenum with only minimal amounts of air in the lower gastro-intestinal tract.

The past history states that the baby's mother had one previous uneventful pregnancy, was Rh positive, and had negative serology for syphilis. This pregnancy was complicated by a threatened abortion at three months. The baby was born after 38 to 40 weeks gestation and weighed 4 pounds 4 ounces (2.04 kg.) at birth. Physical examination at birth revealed no abnormalities, but on the second day of life, a grade 3 harsh, high pitched, systolic murmur was heard over the entire precordium, loudest along the left sternal border at the third and fourth intercostal spaces.

On admission to this hospital physical examination showed a moderately dehydrated, active infant, with temperature 99.8° and weight 3 pounds 15 ounces (1.89 kg.). The abdomen was normal and the aforementioned murmur was present. Urinalysis and blood count were normal. X-ray examination of the chest was reported as a right upper lobe atelectasis. An electrocardiogram showed right axis deviation, which was considered to be within normal limits. The diagnosis of partial duodenal obstruction was entertained, possibly on a neurogenic basis or due to a mucous plug, with anatomic lesions to be ruled out. Because of the absence of cyanosis and cardiac

dysfunction, I-V septal defect or patent ductus arteriosus were considered as likely causes for the murmur.

Treatment was begun with clyses and a dilute skim milk formula in small quantity. No vomiting occurred during the first three days, but as the strength of the formula was increased, vomiting recurred. She continued to have stools and her weight remained fairly stable. The formula was decreased, she was put on antispasmotic medication, and again there was no vomiting for about three days until the concentration and volume of the formula were increased. With the change of the formula from skim milk to olac, vomiting recurred as the concentration of the formula was increased. Throughout this period there were stools every 1–2 days, and her weight showed a slow decline in spite of gavage feedings and a return to skim milk.

By August 8, 1954 her weight had dropped quickly to 3 pounds 6½ ounces (1.63 kg.) and she became quite dehydrated, after which all oral feedings were stopped and she was maintained on intravenous fluids, which included electrolytes, protein, and vitamins. Two blood transfusions were also given. On this routine the dehydration became less marked and her general condition showed considerable improvement, but on several occasions peristaltic waves were seen traveling from the left upper quadrant to the right side of the abdomen. A barium swallow again showed obstruction beyond the first part of the duodenum. Because of the continued obstruction an exploratory laparotomy under ether-O_2 anesthesia was performed on August 12, at which time her general status seemed favorable for surgery. Before operation a tube was inserted into the stomach and 20 ml. of greenish fluid aspirated, the tube being left in place. After the abdomen had been opened, the entire gastro-intestinal tract was explored and marked dilatation of the stomach and first part of the duodenum was noted. Just distal to the dilatation was a narrow band of pancreatic tissue which encircled the second part of the duodenum. A side-to-side duodeno-jejunostomy was done, following which air and sterile water were seen to pass the anastomosis without difficulty.

Post-operatively she reacted completely in about 8 hours, and in 18 hours had active bowel sounds. However, since aspiration of the stomach through the indwelling tube produced 5–20 ml. of fluid every 2–4 hours, she was maintained on intravenous fluids. In 24 hours she had a small bowel movement, but continued to accumulate fluid in her stomach. Her lungs remained clear and there was no change in her heart sounds or the murmur.

About 60 hours after surgery, she suddenly developed slow gasping respirations, but improved after her upper airway was aspirated and she was given caffeine-sodium benzoate subcutaneously. During the next 8 hours she had intermittent apnea and two episodes of cyanosis which were followed by death.

At autopsy poor healing of the anastomosis was noted, which was still held intact by the sutures. The narrow band of pancreatic tissue was seen to encircle a stenotic second portion of the duodenum. Examination of the heart showed a tetralogy of Fallot with associated patent ductus arteriosus and patent foramen ovale. The lungs showed patchy atelectasis, but there was no evidence of aspiration of foreign material.

DISCUSSION

Because this baby clinically and roentgenologically showed the picture of partial duodenal obstruction, conservative management was attempted at first, with a view toward letting her grow larger before a more definite approach was attempted. Furthermore, because of the difficulty of evaluat-

ing the cardiac status it was felt that a delay in surgery would be advantageous. However, in the face of repeated bouts of obstruction, it soon became obvious that regardless of cardiac status, surgery held the only possibility for relieving her symptoms.

Pre-operatively, on intravenous fluids estimated at about 200 ml/kg. per 24 hours, she quickly regained and maintained good hydration. Her daily fluids consisted of one-third electrolyte solution (usually Butler's) one-third invert sugar with vitamins B and C, and one-third protein solution (either plasma or plasma hydrolysate). On this regime she received almost 77 calories/kg. per 24 hours and remained fairly vigorous and alert. She withstood anesthesia and surgery very well and on a reduced fluid and electrolyte intake post-operatively did well until the third day after surgery, when her condition rapidly deteriorated.

Her demise was apparently related to her prematurity, the stress of surgery, and probably her abnormal cardiac function. This case certainly illustrates the importance of thorough abdominal exploration and the disadvantage of merely cutting the pancreatic annulus. It is interesting to speculate whether her ductus arteriosus eventually would have obliterated, changing the picture from acyanotic to severe cyanotic heart disease.

In conclusion we would feel that intestinal obstruction, even if partial, in the premature, as in any other age group, is an indication for surgery as early as possible.

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